289

Colonoscopy in adult cystic fibrosis: A 24-year experience in a single CF centre

H. Chaun[1], D. Owen[2], P. Zetler[2]
[1]*Department of Medicine (Gastroenterology & Adult CF Clinic),* [2]*Department of Pathology, University of British Columbia, Vancouver, B.C., Canada*

Intestinal symptoms are common in cystic fibrosis (CF) patients, and abdominal pain is a significant cause of morbidity. AIM of this study is to review the indications and the frequency of abnormal findings in adult CF patients investigated with colonoscopy and biopsies. METHODS: Since 1980, 112 patients have been referred to Gastroenterology from the Adult CF Clinic. A retrospective chart review showed 60 colonoscopies were performed in 30 patients, single colonoscopy in 18, 2 in 8, multiple in 4 patients. 20 were females, 10 males, age range19-57 years (mean 29.5). Primary indication was abdominal pain 16, bleeding or anemia 7, diarrhea 3, Crohn's disease 2, previous colonic cancer 1, previous sigmoid resection and colostomy 1, bowel dysfunction 1, abnormal barium enema 1. RESULTS: 14 patients had abnormal colonoscopy – acute segmental colitis 2, proctitis 1, Crohn's 2 (both with granulomata), diverticulosis 1, previous resection for carcinoma 1, polyps 4 (adenoma 3, juvenile 1), obstructive colonic carcinoma 1, enlarged appendix with impacted fecolith 1, DIOS 1. 24 patients had complete colonoscopy (to terminal ileum or cecum) and biopsies – 14 had right-sided microscopic colitis, including 10 with abdominal pain. There were no complications. CONCLUSION: 20/30 patients (66.6%) had abnormal colonoscopic and/or biopsy findings. Most importantly, carcinoma was diagnosed in 1, another had previous resection for colonic cancer; 3 had tubular or tubulovillous adenoma; 2 had Crohn's disease. 14 had right-sided microscopic colitis, clinical significance undetermined, 10 of whom had abdominal pain. Colonoscopy is a safe and essential investigation in adult CF patients with obscure intestinal symptoms.

290

Liver disease in children with cystic fibrosis: prevalence and clinical characteristics

M.G. Slieker, H.P.J. van der Doef, C.K. van der Ent, R.H.J. Houwen
CF Center Utrecht, UMC Utrecht, Utrecht, Netherlands

Aims*:* CF related liver disease (LD) has been universally associated with a severe genotype. Some studies described other associations, the most recent being more severe lung disease and a general growth failure. In this study we studied the prevalence of LD in patients with CF and identified clinical characteristics associated with the presence or absence of LD.
Methods*:* The prevalence of LD was calculated using the results of the last yearly multidisciplinary check-up. LD was defined as the presence of at least 2 of the following conditions: hepatomegaly, abnormal serum liver enzyme levels and ultrasound abnormalities other than hepatomegaly. A case-control study was conducted to identify clinical characteristics associated with LD. Each child was pair matched for sex, date of birth and age with a CF patient without evidence of LD.
Results*:* LD was found in 52 of 238 children with CF (22%) aged between 5 and 19 years. Male gender was more common in patients with LD (58% vs 48%, P=0.21). The presence of LD was associated with a severe CFTR genotype (P<0.001) and with lower height for age (P<0.01). No association between LD and a history of meconium ileus, weight or pulmonary function (FEV_1, FEV_1/FVC, RV/TLC and TLC) was found. Analyses of subgroups with severe hepatomegaly or liver protein synthesis impairment (factor V<60%) did not allow us to find additional associations, although lower height for age was more often observed in the latter subgroup (P=0.02).
Conclusions*:* The prevalence of LD in children with CF is high (22%). CF patients with LD have more frequently a severe genotype and impaired growth. Contrary to some recent studies, in our population the presence of LD was not associated with male gender, a history of meconium ileus, pulmonary function or weight.

291

The prevalence and risk factors for the development of cystic fibrosis related liver disease

S. Fustik[1], M. Trajkovska[1], N. Slaveska[1], T. Josifovska[1], S. Koceva[2], G. Efremov[2]
[1]*Clinical Centre, Skopje,* [2]*RCGEB, MASA, Skopje, R. Macedonia*

Liver disease (LD) in cystic fibrosis (CF) has become a matter of greater concern since the population of CF patients lives longer. The aim of this study was to define the prevalence and the role of possible clinical and genetic risk factors for the development of CF- related LD. All patients older than 3 years (n=52) followed at the CF Centre in Skopje were screening for LD through clinical, biochemical, ehographic, and hepatobiliary scintigraphic assessment. LD was defined by the finding of hepatomegaly and/or splenomegaly; significant and persistent increased of at least two serum liver enzyme levels; suggestive ultrasonographic abnormalities (score >4); and morphologic or functional scintigraphic abnormalities. According to our predefined criteria, 18 patients (34.6%) were classified as having LD, three of them with portal hypertension. Clinical presentations of LD had 6 (33.3%), abnormal liver blood test results 12 (66.6%), ultrasound abnormalities 18 (100%), and scintigraphic abnormalities 13 (72.2%) of the patients with LD. Prevalence of LD increased with age (average 13.2±5.3 years in the LD group vs 9.9±5.6 years in the no LD group). A male predominance was found in the group with LD (72%). Pancreatic insufficiency was present in all patients with LD There was no significant difference in the pulmonary function, nutritional status, and in the prevalence of meconium ileus or DIOS. Genetic analysis show higher frequency of ΔF508 mutation in LD group (77.8%) vs no LD group (66.2%). All patients with LD had severe mutations: ΔF508, G542X, N1303K, CFTRdel.21Kb, 1811+1G→C, and Y1092X. In conclusion, male and older CF patients who have pancreatic insufficiency and carry mutations associated with a severe phenotype are in increased risk to develop LD.

292

Long-term effect of ursodeoxycholic acid on the Cystic Fibrosis related Liver Disease

M. Fotoulaki[1], E. Kotsi[1], K. Vassilaki[1], A. Galli-Tsinopoulou[1], E. Pavlitou[2], S. Nousia-Arvanitakis[1]
[1]*Fourth Department of Pediatrics, Aristotle University, Thessaloniki ,GR,* [2]*Immunology Lab, Papageorgiou General Hospital, Thessaloniki, GR*

We have demonstrated that ursodeoxycholic acid (UCDA) improved cholestasis and hepatic function in nodular biliary cirrhosis (NBC) and also, reversed the early sonographic findings of focal biliary cirrhosis (FBC) during a 10 year- follow up period (1990-2000) .The **aim** of the present study was to further evaluate the effects of UCDA on the natural history of NBC in the following period of 4 years (2001-2004). **Materials and methods**: 7 CF individuals, aged 12 to 29 years, who had NBC, had been followed-up regularly and had excellent compliance, underwent detailed physical examination, liver function tests and ultrasound liver scan at six-month intervals. Hormone profile (estradiol, androstenedione, sex hormone binding globulin and prolactin) was measured. The results were compared with those of three CF patients aged 12-16 years, who had irregular follow-up and no compliance to the physician's instructions. **Results:** Liver and spleen size remained stable, liver function tests as well as full blood counts were normal, sonographic findings did not deteriorate and there was no variceal bleeding in the 7 compliant patients. Hormone profile was normal in these patients indicating normal liver function. By contrast, the three noncompliant CF-NBC patients demonstrated abnormal liver function tests; deteriorating portal hypertension with hypersplenism and minor episodes of hematemesis they also had increased levels of prolactin. **Conclusion**: USDA administration appears to have a positive influence on the progression of NBC in CF patients.